SACHS (B.) & ARMSTRONG (S.T.)

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In 1883 Dr. Morvan, residing in a little town in Brittany, published a paper on a disease that he had observed there to which he gave the name of analgesic paresis and panaritium of the superior extremities, or pareso-analgesia. In this paper he stated that at the commencement the disease was limited to one extremity, subsequently passing to the other, and always terminating in the production of one or more felons.

In the cases as first described by Morvan the symptoms are initiated by a weakness of the muscles, and sometimes by a pain in the forearm, that is succeeded by a swelling of the member, with the formation of deep palmar fissures and felons, usually painless, with phalangeal necrosis. It was for the latter condition that the physician was consulted, and at that period there was usually paresis of the muscles of the affected region that was afterward followed by atrophy of the thenar, hypothenar, and interosseous muscles. While faradization would produce energetic contractions in the muscles of the forearm, no reaction would

be obtained in the atrophied muscles. There was analgesia of the forearm, sometimes of the arm, neck, and chest; and also thermal, but no other anæsthesia. Exertion produced occasionally hyperidrosis of the analgesic region; and the existence of vaso-motor disturbance was further evidenced by the bluish or mottled discoloration of the affected part in cold weather, and the occasional formation of phlyctænulæ. Of his reported cases, seven were in males and two in females, and the disease had lasted from a few to twenty-five years without involvement of other regions.

It seems to us that the report that Morvan made of what seemed to him to be a new disease should be considered in giving it a place in nosology. The existence of paresis, loss of pain sense and thermal sense, circumscribed atrophy of the forearm or hand muscles, and trophic disturbances evidenced by the formation of cutaneous fissures and felons, would constitute what is known as Morvan's disease. In all reported cases of the disease made at a subsequent date these have been the essential symptoms, and the following case is added to the literature of the subject, the patient having been presented for examination to two medical societies of this city:

W. H., aged twenty-eight, a native of Germany, a laborer, was referred to Dr. Sachs's clinic at the New York Polyclinic by Dr. Gerster. The patient had a venereal ulcer and a bubo in the right groin ten years ago, but otherwise he has always been healthy until four years ago, when he was employed as a dish-washer, his hands lost their muscular power, there was slight twitching and enlargement of the fingers, and the skin of the fingers and hands became thickened and fissured. He was treated by a physician, and the enlargement in the fingers subsided, except in the index and third finger of the left hand, in which felons formed, that were incised without causing any pain. He thought that the condition of his hands resulted from the use of soda in the wash-water; but it is now four years

since he stopped washing dishes, and his hands have not improved. At the time of examination the skin of each hand was of a purplish color, that was intensified by cold weather, the discoloration under such influence extending up the arm. The hands themselves presented the following appearance:

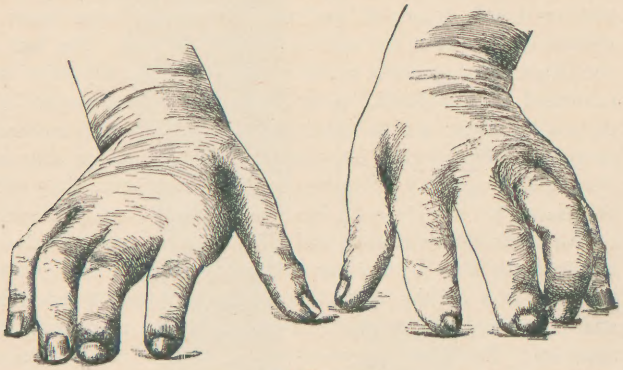

FIG. 1.

Right. The skin on the dorsum seems to be normal, but on the dorsum of the fingers it is thickened; there is a slight contracture at the second phalangeal joint in all of the fingers, but more pronounced in the middle and index fingers. There is a small eschar on the dorsum of the thumb, but this member is not contractured. There is a marked atrophy of the first dorsal interosseous muscle. On the palmar surface the skin is thickened, and there are numerous ragged excoriations, especially on the finger-tips and at the base of the middle and ring fingers; in these excoriations deep fissures, having indurated edges, have formed. On the anterior surface of the right forearm is an area of dermatitis resembling a mild degree of ichthyosis. At the bend of the elbow there is an area four inches long by an inch wide, in which there are numerous small depressed atrophic areas that might be described as a *dermatrophia circumscripta albida;* the patient thought that this had resulted from carrying a basket on his arm, the markings resembling somewhat

those that would be produced by the pressure of the twisted willow in the handle thereof. There is a scar over the olecranon, caused by an incision (painful) for an abscess in 1890. Left hand: The skin on the dorsum and palmar surfaces of the hand and fingers presents a similar appearance to that of the right hand, but the nails of the index and middle fingers are thickened and deformed, and the end of the index finger is conical while that of the middle finger is clubbed. The nail of the latter finger presents white opaque striæ, and a portion was examined microscopically to see if these striæ were caused by a mycelial growth; but no fungus was found. There is moderate atrophy of first dorsal interosseous. Dynamometer showed: *Manus dextra*, thirty kilogrammes; *manus sinistra*, sixty kilogrammes; but this disparity has been lessened during the course of the electrical treatment, and the muscular power is almost equal at the time of writing this report, though it is yet less than that of a healthy man. The muscular sense was normal, and, excepting in those muscles above mentioned, no atrophy was apparent. The forearms were well developed and were equal in circumference. The tactile and pressure senses were normal, the patient locating a straw drawn over the skin, and discriminating between different weights. The pain sense is abolished in an area on the dorsum of the right hand; also over the dorsum of the fingers, hand, and ulnar side of the left forearm; but there is no loss of pain sense in the palms or the anterior surface of either forearm. Fig. 2 shows the analgesic areas on the hands; the test was made by forcing a needle into the flesh. The temperature sense did not recognize a temperature of 212° F. on either forearm, excepting at the flexure of the elbow, where it felt hot; but at this point a temperature of 150° F. felt cold. Temperatures of 190° to 200° F. were recognized as warm on the upper portion of the arms and back, though lower temperatures were called cold. In the regions above mentioned a temperature of 32° F. was not recognized as very cold, even if it was placed on a spot on which a temperature of 212° F. had just been placed. These observations were made with test tubes containing boiling water and a freezing mixture. There is fibrillary twitching of the muscles of

the forearm and hand. Fig. 1 shows the atrophy of the interossei, the deformity of the fingers of the left hand, and the slight

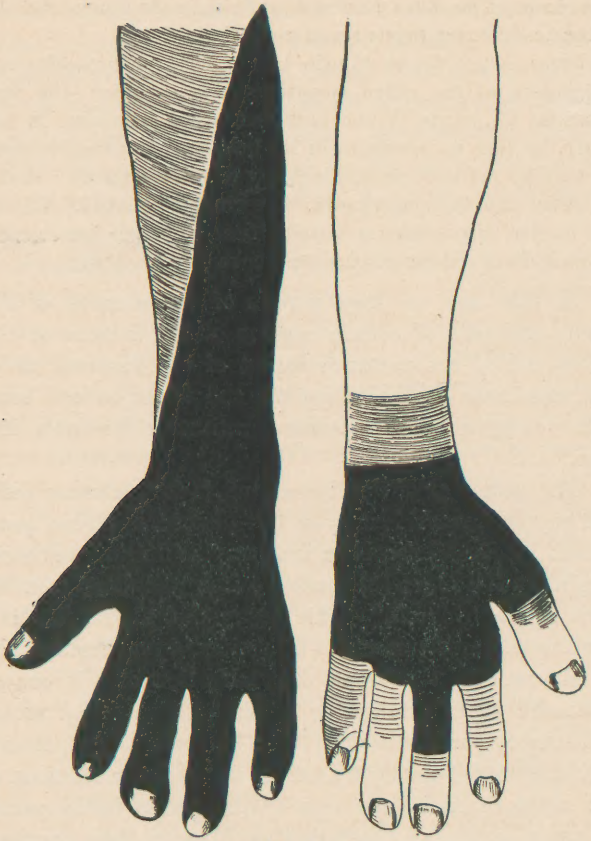

FIG. 2.

tendency to the *main en griffe*. An electrical examination showed an absence of the faradaic reactions in the extensor, thenar, and interossei muscles, though the flexor group reacted

well. Galvanic reactions, ACC>KCC in the extensor muscles of each forearm and the interossei. It might be here stated that since the treatment by electricity has been commenced, the faradaic reactions have returned in the extensor groups and the difference in the galvanic reactions is diminishing.

There is no history of pain in the forearms preceding the appearance of the other symptoms; his attention was first called to his hands by the fact that, when immersed in hot water, he had no sensation of its heat—a phenomenon associated with the swelling and inability to use his hands. When his entire skin is perspiring his forearms will be cold. He has had to give up positions as a waiter because he has been unable to firmly hold articles of glass or crockery in his hands.

In this case we have paresis of the muscles of the hand; analgesia of certain regions of the dorsum of the hands and of the posterior aspect of one forearm; thermal anæsthesia of both forearms, and felons on one hand —the congeries of symptoms constituting Morvan's disease.

The pathology of the disease, and, in fact, whether there was any such morbid entity as this disease, has been questioned. Osler (2) considers it a peripheral neuritis of toxic origin, and Gowers (3) considers it a peripheral neuritis with myelosyringosis.* This theory that the disease is a peripheral neuritis seems to be verified by an examination of Morvan's first reported cases; his first case is one of traumatic neuritis following a fall, with persistent motor and sensory paresis for ten years; his second case presented symptoms of multiple neuritis, in which the affection of the nerves of the lower extremity disappeared in the course of years, certainly a result that would never have occurred in myelosyringosis; his third case seems to be one of

* Our attention has been called to the barbarism in the composition of syringomyelia, and we have adopted myelosyringosis as a term that avoids the etymological error in the more usual word.

chronic neuritis, as is evidenced by the occurrence of painful paroxysms during twenty-four years; his fifth case resembles traumatic neuritis; and his seventh case resembles one of multiple neuritis. Monod and Reboul, in their report of a case of the disease, took the position that it was a variety of peripheral neuritis, having found in an examination of the nerves of an amputated finger an acute parenchymatous and interstitial neuritis. Déjérine (13) thought that the frequent appearance of the disease in the population of a small province showed that it was a neuritis of toxic or infectious origin.

Surgical pathology teaches us that felons do not originate spontaneously, but in consequence of the introduction of the *Streptococcus pyogenes;* and the fact that the felons are usually painless seems to show that the micro-organism gains access to the tissues in consequence of the trophic disturbances, and that the felons are merely an incident that might be prevented in such cases by due attention to cleanliness.

The dissociation of sensory symptoms is the chief argument against the theory that Morvan's disease is due to peripheral neuritis, but there is evidence that all sensations are not equally affected in neuritis, as has been asserted by Starr (5); and possibly the paucity of such records is due to failure to make special tests.

As it is a physiological fact that the conduction channels of the tactile, pain, and thermal senses lie in different parts of the spinal cord, it seems a justifiable assumption that their peripheral terminations are also different. And that recorded cases justify this assumption is evidenced by the cases of Weir Mitchell (6), in which there was a lessened sense of pain with no loss of touch; those of Gowers, in which he has seen loss of pain sense while the muscular sense is preserved; and those of Grainger Stewart (7), in

one of which the thermal sense was diminished while the pain and muscular senses were normal, and in another the thermal and pain senses were diminished and the muscular sense was normal. The latter case is especially serviceable in supporting the possibility of the existence of such phenomena in neuritis, because the necropsy showed that in the median, ulnar, and tibial nerves certain "bundles of nerve fibers were totally, others partially destroyed, while some were comparatively healthy"; and in the cervical enlargement of the spinal cord there were tracts of secondary degeneration, affecting only the columns of Goll and the outermost part of the lateral columns.

These cord degenerations were in consequence of an ascending neuritis, and an explanation of the gliomatosis of the cord, in cases of Morvan's disease, is possible on the ground that there was an ascending neuritis of the sensory fibers, with later slow gliomatous degeneration in their tracts in the spinal cord. Gombault (8) found in a necropsy, in a patient who had Morvan's disease for forty-four years, intense changes in the peripheral nerves, with a mild degree of sclerosis of the posterior horns and columns. These facts justify the statement of Gowers regarding this disease, that "we must be cautious in inferring that the pathological state is the same in origin in all cases." But physiological, pathological, and clinical data support the idea that a peripheral neuritis may be the cause of the disease, though Morvan himself considers it is of spinal origin.

Myelosyringosis so closely resembles Morvan's disease in its early stages that several prominent neurologists—such as Bernhardt, Jolly, and Charcot—have considered them identical; and the latter proposed that the congeries of symptoms constituting the former disease should be denominated myelosyringosis of Morvan's type. Now, myelosyringosis is purely pathological condition that may include, according

to Cheron (10), first, dilatation of the central canal, or myelohydrosis; second, the excavating myelitis of Joffroy and Charcot; third, the peri-ependymal sclerosis of Hallopeau; and, fourth, gliomatosis of the region of the central canal. And with these various pathological conditions Joffroy and Achard concluded (11), from a study of the disease in general, that often a sufficient number of the supposedly pathognomonic signs are not present to allow a diagnosis to be made; again, that where all of these signs are present they may suddenly disappear, and a spontaneous recovery is hardly to be expected in such a disease—in other words, a peripheral neuritis has been mistaken for myelosyringosis. In the following table we present a comparison of the essential features of both diseases:

| *Myelosyringosis.* | *Morvan's Disease.* |
|---|---|
| Felons rarely present, and only as a symptom of a trophic disturbance. | Formation of painless felons. |
| Fissures rare. | Palmar cutaneous fissures. |
| Analgesia of areas supplied by the segment of the affected cord; usually arms and upper half of trunk; rarer in lower part of trunk and legs. | Analgesia of fingers, of hand, and forearm; later and rarely of arm and neck. |
| Thermal anæsthesia of analgesic and other regions; sometimes unequal for heat and cold, sometimes perverted. | Thermal anæsthesia extending moderately beyond the analgesic areas. |
| Muscular atrophy of region supplied by nerves emerging at or immediately | Muscular atrophy usually limited to thenar, hypothenar, and interosseous re- |

below the level of the affected segment of the cord.

Tactile sense sometimes lost.

Often neuralgic pains in joints of the affected region and in the spine.

Occasional Romberg symptom. Unsteadiness of movements. Paralysis of one vocal cord; of tongue or face. Dysphagia. Dyspnœa. Cardiac irregularity. Inequality of pupils. Occasional nystagmus and ptosis. Occasional spastic paralysis of lower limbs.

Bones may become thick and brittle and tabetiform joint changes may occur.

Bilateral in eighty per cent.

*Mains de prédicateur* in consequence of predominance of extensor paralysis [Morvan].

Symptoms usually develop slowly, increasing gradually in the course of years; death from exhaustion or impairment of function. Recovery rare.

gions; more rarely the forearm.

Tactile sense normal.

Pain may precede the other symptoms, rarely persist.

No Romberg symptom.

Rarely any affection of bones (excepting necrosis from felon) or joints.

Bilateral in forty-five per cent.

*Main en griffe* in consequence of predominance of flexor paralysis.

Symptoms usually develop rapidly; most often confined to the forearm and hand. No extension of symptoms in from ten to forty years. Recovery, or at least marked improvement, not infrequent.

Joffroy and Achard (14) reported a necropsy made on a woman who had, forty-five years before her death, *painful* felons of both hands, leaving deformities of the fingers resembling Morvan's disease; sensibility both to pain and heat was, just before her death, greatly diminished in the palmar surface of her hands and fingers, and the tactile sensibility was diminished; but there was no muscular atrophy and the electrical reactions were normal. She also had kyphosis. At the necropsy not only was a cavity found in the spinal cord, but the nerves of the forearms had undergone extensive degenerations; these latter they regard as secondary to the lesion in the spinal cord, just as is the peripheral neuritis that has been observed in posterior spinal sclerosis. It might be questioned whether, excepting in physical appearance, this case presented any similarity to Morvan's disease. The felons were painful; the deformities that followed them did not interfere with the usefulness of the hands; the muscles were not atrophied; the electrical reactions were normal; and the sensory disturbances occurred at the age of seventy-five, when sensation would naturally be rather sluggish, and even then a temperature of 140° F. was recognized as something warm, and the pin prick was indistinctly felt in the thickened skin of the palm and fingers.

We do not desire to maintain that cases of myelosyringosis do not present symptoms in the earlier stages of the disease closely simulating all the phenomena of Morvan's disease; and it is furthermore probable, as Joffroy and Achard have stated, that lesions in the bulb may produce such symptoms, and that supposed cases of Morvan's disease have really been cases of myelosyringosis. But we would maintain that cases of the latter disease of sufficiently long standing will present later and more serious complications than those reported as characteristic of the former malady.

That it is not necessary to found the existence of the disease on a pathological condition in the spinal cord is, we think, demonstrated by the observations of Charcot, who has found sensory dissociation in hysteria just as it is observed in the disease under consideration, and myopathic phenomena also. These would leave the felons as the single absent symptom, and we know their presence is due to a definite cause. Minor, of Moscow, has observed sensory dissociation in traumatic myelæmatoma; and the presence of this dissociation is so frequent in anæsthetic leprosy that the possibility of the identity of that and Morvan's disease has been broached.

For instance, Dr. Zambaco (12) has held that both myelosyringosis and Morvan's disease are identical, and that the disease is only a form of anæsthetic leprosy that has been, so to speak, attenuated by the manners and climate of Europe; this has been warmly controverted by Thibierge (16), and one of us can personally state that there is no resemblance between the present case or those reported cases that have been consulted and the cases of anæsthetic leprosy he has observed in the southern American states and in Norwegian leper hospitals.

We believe that Raynaud's disease, erythromelalgia, and sclerodactylia are sufficiently typical not to be confused with Morvan's disease.

While, therefore, it is to be distinguished from myelosyringosis, hysteria, and, in leprous countries, from anæsthetic leprosy, we believe the presence of other phenomena in those cases will enable a diagnosis to be made.

From the preceding presentation of facts, and our own experience with myelosyringosis and neuritis, we infer that the typical cases of Morvan's disease may be due to a peripheral neuritis. On the other hand, we have conceded that cases of myelosyringosis may so closely resemble Morvan's

disease as to make a distinction impossible; but the strict limitation of the disease usually for many years and the early appearance of the painless felons, as well as the improvement, if not recovery, in given cases would weigh strongly in favor of the diagnosis of Morvan's disease rather than myelosyringosis. And, lastly, it must be conceded that it is possible for an ascending neuritis to lead to gliomatous degeneration in the central canal of the spinal cord, such cases presenting later the typical clinical features of myelosyringosis.

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